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case report

# Primary urachal adenocarcinoma: a rare case report

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## Abstract

Primary urachal carcinoma is a very rare cancer with a poor prognosis. It generally presents as a high-grade, high-stage tumor, and in most cases the patient has developed regional or distant metastasis at the time of presentation. Here, we report a very interesting case of primary urachal adenocarcinoma with signet ring cell carcinoma in a 58-year-old male who presented with a lower abdominal mass and discomfort. In this case, urachal carcinoma was successfully treated with surgery using an extended partial bladder cystectomy approach with excision of the urachal mass and umbilicus. The patient also underwent systematic chemotherapy with 5-fluorourical and cisplatin. During the 12-month follow-up period, the patient did not experience recurrence or metastasis. Overall, we found that an organ preserving extended partial cystectomy along with chemotherapy was an optimal treatment method that helped improve the patient's quality of the life with no recurrence of cancer so far.

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The urachus is a tubular structure that connects the bladder with the umbilicus. Nitrogenous waste is discharged from the bladder, the main organ of excretion during the fetal period. During the fourth and fifth month of embryonic life, the urachus gradually degenerates into a rudimentary fibromuscular closed canal, which is known in adults as the median umbilical ligament, and stretches between the dome of the urinary bladder and the umbilicus. However, a urachal remnant in the form of a tubular or cystic muscular structure can persist. It is most commonly found between the umbilicus and the bladder, usually at the dome of the bladder. After birth, if the umbilical lumen is not completely closed, it may lead to various abnormalities including infection, cystic degeneration, and malignancy [1]. Autopsy studies suggest that in one-third of adults, the urachus canal partly persists [2]. Urachal carcinoma (URC) is a rare malignant disease that manifests in residual urinary tissue, accounts for 0.01% of adult tumors and approximately 20% to 40% of primary bladder adenocarcinomas [3]. Urachal tumors tend to be associated with a poor prognosis, with 5-year survival rates ranging from 9.0% to 43% [4].

## Case report

A 58-year-old male, with no smoking history, presented to us with a lower abdominal lump approximately 5.0 × 3.0 cm in size. The lump had started to develop five years prior and grew gradually each year. As the lump grew, he also felt discomfort. He had no previous history of hematuria, cystitis cystica, cystitis glandularis, bladder irritation or weight loss. However, the patient had a 24-year history of hypertension for which he took medication.

On physical examination, a well-defined lump was palpable in the umbilical and hyponastic region and was globular in shape. It was approximately 12 × 10 cm in size, had smooth surface and regular margins all around. No abnormalities were found in the rest of the physical examination. Ultrasonography showed a solid

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and cystic heterogenous, uneven mass approximately 12.4 × 10.1 × 9.2 cm above and close to the left wall of the bladder. The ultrasound also showed evidence of adequate bladder filling, smooth mucosa and no obvious significant mass. The urine microscopy and urine cytology did not reveal any abnormalities. Computed Tomography (CT) of the abdomen showed a hypodense soft tissue lesion in the supravesical region near the dome of the urinary bladder; the lesion extended superiorly up to just below the umbilicus, and therefore the border between the lower margin and the bladder was not clear (Fig. 1). There was no evidence of distant metastasis or intra-abdominal lymphadenopathy and the other organs appeared normal. The provisional diagnosis was a urachal

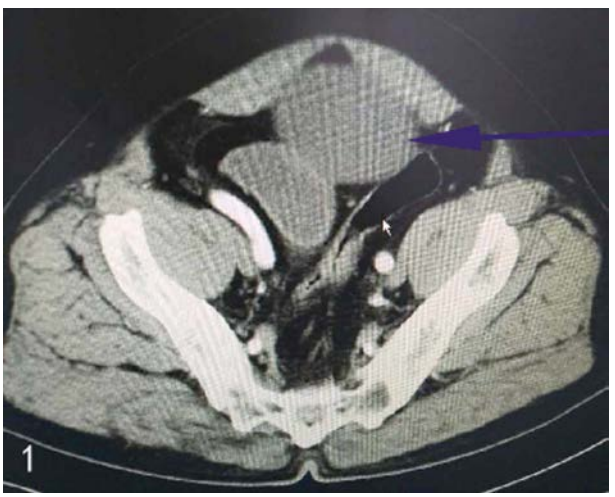


**Fig. 1** Computed Tomography of abdomen showing urachal mass (arrow) attaching the bladder dome and squeezed the bladder

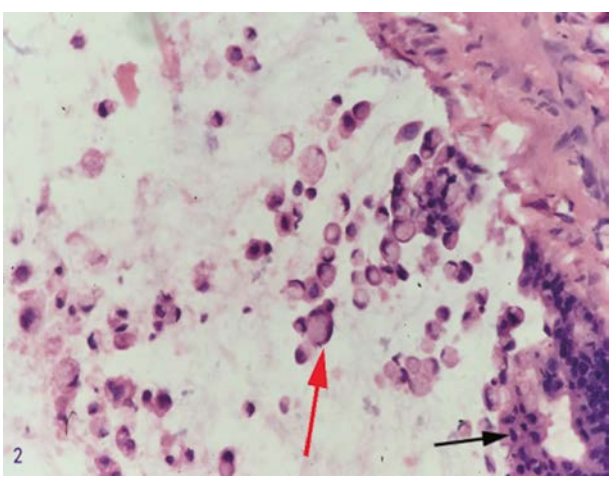


**Fig. 2** Histopathological findings of the biopsy showing the tumour cells containing intracellular vacuole displacing the hyperchromatic nucleus to one side suggestive of signet ring cell carcinoma (red arrow), l urachal mucinous adenocarcinoma (black arrow)

remnant malignancy. The patient underwent cystoscopic examination, which revealed a round swollen area of tissue that was squeezing the anterior dome of the bladder; the bladder mucosa was smooth.

Surgical exploration was performed for the patient after adequate preoperative preparation. During the exploration, a urachal mass extending up to the dome of urinary bladder was found. The mass was approximately 12.0 × 10.0 × 9.0 cm in size and was not interlinked with the bladder. Surrounding it was approximately 200 milliliters of a viscous secretion. An extended partial cystectomy with excision of the urachal mass and umbilicus was performed after confirming the absence of leakage from the urinary bladder. After the operation, the patient received 6 cycles of systemic chemotherapy which included 5-flurourical (FU) and cisplatin.

Pathological analysis revealed a urachal mucinous adenocarcinoma, with partial signet ring cell carcinoma; there was no evidence of deep invasion through the urachus, and the incisal margin of the bladder was negative (Fig. 2). The Myao stage at diagnosis is I period. The patient has been regularly followed-up for 12 months and doing is well.

## Discussion

Primary urachal adenocarcinoma is an extremely rare but highly malignant tumor, accounting for only 0.17 to 0.34 % of all bladder tumors [5]. This tumor is more common in males aged from 40 to 60 years. To date, no consensus has been reached regarding the diagnostic criteria of URC. The most commonly used criteria have been proposed by Sheldon *et al* [6] and Mostofi *et al* [7] and revised by Gopalan *et al* [5] which include the following characteristics: (1) tumor is located in the dome/anterior wall of the bladder, (2) the tumor is located at the epicenter in the bladder wall, (3) there is an absence of cystitis cystica and cystitis glandularis, and (4) lack of known primary adenocarcinoma elsewhere. The most common symptoms of URC include macroscopic or microscopic hematuria, abdominal pain, and dysuria. Other less common clinical presentations included pollakisuria, pyuria, urinary tract infection, umbilical discharge (e.g. blood, urine, and mucus), vaginal discharge, and nonspecific symptoms (nausea, vomiting, diarrhea, weight loss, or fever) [8]. Some serum markers have proven to be helpful in the diagnosis and monitoring of URC, including carcinoembryonic antigen and carbohydrate antigen 19-9 as well as cancer antigen 125. Increased serum levels of these markers have been detected in patients with URC adenocarcinoma[9, 10]. In this case, the patient presented with a mass in the abdomen, no hematuria or abdominal pain, absent typical clinical symptoms such as___ and the serum markers appeared normal. When a patient presents with non-

specific symptoms like an abdominal mass in our case, then a high index of suspicion is required because these symptoms are very common with benign conditions. The most common pathological type of urachal tumor is adenocarcinoma. Other rare patterns include the signet ring cell type, clear cell type, hepatoid type, and mixed patterns [11]. It has been postulated that the oncogenesis of urachal adenocarcinoma involves a metaplastic process, as the urachal urothelium often exhibits glandular metaplasia [5]. Several stage classifications exist, but the most often used is the Mayo staging system [6], which includes the following: I. Tumor is confined to the urachus and/or bladder, II. Tumor extends beyond the muscular layer of urachus and/or the bladder, III. Tumor infiltrates the regional lymph node, IV. Tumor infiltrates the non-regional lymph nodes or other distant sites. Generally, URC presents as a high-grade, high-stage cancer, and in the majority of cases there is regional or distant metastasis at the time of presentation. Therefore, this cancer is usually associated with a poor prognosis [12]. By lymphatic dissemination URC usually metastasizes into the pelvic lymph nodes and by hematogenous dissemination into distant organs, especially lungs, bone, or peritoneum [13]. Although the pathological features of ureteral adenocarcinoma and bladder adenocarcinoma are similar, the prognosis for ureteral adenocarcinoma is better than for bladder adenocarcinoma. This may be due to the age of patients since those with ureteral adenocarcinoma are often younger than those with adenocarcinoma of the bladder [14]. Surgery remains the primary treatment for prolonging the overall survival of patients. Some scholars believe that both partial and radical cystectomy can be considered as they provide similar oncological results [15]. However, Behrendt *et al* [16] suggests that an organ preserving extended partial cystectomy provides a higher quality of life and should be preferred. Since URC is not sensitive to radiotherapy, chemotherapy, along with the surgery, is the only treatment option to potentially prolong survival. One meta-analysis suggested that the most effective treatment may be a combination of 5-FU with cisplatin, which performs significantly better than cisplatin-based only therapies [8].

URC is an extremely rare cancer and often presents as a high-grade, high-stage cancer. A localized URC organ preserving partial cystectomy provides a long-term disease-free survival, and the combination of 5-FU with cisplatin provides the most favorable response. In addition, regular postoperative follow-up is necessary.

### Conflicts of interest

The authors indicated no potential conflicts of interest.

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